Modern Concepts of Cardiovascular Disease

Published monthly by the AMERICAN HEART ASSOCIATION

1790 BROADWAY AT 58TH ST., NEW YORK, N. Y.

DR. J. HAMILTON CRAWFORD, Brooklyn, *Editor*

DR. CHARLES A. POINDEXTER, New York, *Associate Editor*

Vol. XI **July, 1942** **No. 7**

THE NATURAL HISTORY OF RHEUMATIC CARDIAC DISEASE

Of the two great causes of disability on account of diseases of the heart, senescence and rheumatic fever, rheumatic fever is the second. Its incidence is probably less than a generation ago. Between the ages of 5 and 25 its *annual death rate in New York City* is 8 to 14 times higher than that in the general population and after the age of 25, 3 to 5 times higher. It is not yet certain but there is accumulating evidence to show that in other climates there is a difference from the experience in New York City and the Northeastern part of the country. In the South and Southwest there may be fewer cases. The assumption is that the difference depends partly on climate and partly on the varieties of bacteria which that climate and atmosphere (sunlight) permit. Precisely what the causes of rheumatic fever are is also still not exactly known. No explanation which associates a single bacterial strain with rheumatic fever is now accepted. That certain kinds of streptococci occur with a high degree of uniformity seems clear. But simple infection in the way in which pneumococci are related to pneumonia, or typhoid bacilli to typhoid fever, does not seem to be the case in rheumatic fever. Here patients are in some way made vulnerable to infection either because of preliminary preparation by another agent not yet identified or by preliminary infection with strains of streptococci themselves. After a certain period during which unknown changes take place in the body the manifestations of rheumatic fever become possible.

In this brief report, not an account of the clinical aspects of rheumatic fever is to be described but an account of its natural history. This account is based on a study of 11,109 cases in the files of the Research Committee of the New York Heart Association. After the age of 10, males in this group were more numerous than females. Because the meaning of a sign or of a symptom cannot be appraised so far as prognosis is concerned until death, the statistics which are to be given apply only to 3,129 cases known to be dead.

As is already well known rheumatic cardiac disease is found at all ages, 65% of the 3,129 cases occurring in childhood. Childhood in this case is arbitrarily taken to extend to 12 years in females and 14 years in males. In adolescence, which extends again arbitrarily to 18 years in females and 20 in males, the cases to be found have fallen to 56%, already 9% less than in childhood. This includes the survivors from childhood plus the new cases added during this period. In early maturity, up to 29 years both in men and in women, the number afflicted was 42% in which are included the survivors from childhood and adolescence, plus the new cases. Here there is a drop of 14% from the numbers in adolescence. In middle maturity, up to age 45, there are those cases which have survived to this age plus, of course, the new cases. But of the rheumatic population there are now only 36%. After this age only 16% of the entire number of cases of rheumatic fever alive are to be found. This is only another way of saying that rheumatic fever is to be found preponderantly among children; the maximum number has its beginning at the age of 8. If all the cases of rheumatic fever are taken into account the mean age at onset is 15 both for males and females. The mean ages would naturally be different if they were based on experience either in children's clinics or in clinics of adults. When there is no loading in favor of any age, 15 seems to be the mean. And at that age, 70% of all persons afflicted have already acquired the disease.

The first manifestations of rheumatic fever vary in young and in adult life. In young persons, and there is little difference between girls and boys, polyarthritis is the dominant complaint in about half. Chorea and infection of the heart occur in about a third. As time goes on polyarthritis becomes less important so that after 40 it is seen in not more than a fifth while infection of the heart occurs only rarely. At the same time the valvular lesions of the heart without signs or a history of infection make their appearance in increasing numbers; so that after 40 they account for not less than 80% of rheumatic maladies. It has always been understood that growing pains and chorea are to be found more especially in younger people. In this experience it is striking that after 20 there are practically no such cases. These are the two manifestations which appear more frequently in girls than in boys. A word should be said on muscle and joint pains. They were discarded as a way of dating the onset of illness if they were the sole complaints and no further rheumatic manifestation appeared later. Many details concerning the distribution of symptoms are matters of more than passing interest. Most cases of chorea (92%) occur under the age of 13; muscle and joint pains and carditis (86%) occur before that age. Polyarthritis, on the other hand, falls at a relatively lower level (60%) and valvular disease alone at a lower one still (36% under 13). After 21 polyarthritis still persists as a significant first symptom (20%) and valvular disease alone becomes distinctly important (50%).

Infection of the heart occurs as a first manifestation at a maximum of about 6 years and polyarthritis from 6 to 8. Chorea presents itself in maximum numbers at the age of 8, and muscle and joint pains from 8 to 10. Although these are the ages at which for each of these manifestations the maximum number of cases appears, they are, as significant first symptoms, at an end by 15 or 20 years in muscle and joint pains and at 20 in the case of chorea. As first symptoms polyarthritis and valvular diseases are not crowded together in the early years as are these, but spread out so that in valvular diseases alone they keep occurring in fairly large numbers to fairly advanced ages.

This may be the point at which to pause in order to utter a warning on using statistics for prognostic purposes. What is said in the way of prognosis on the basis of statistics has only indirect meaning in individual cases. Statistics are *average* experience

only. Even if the average duration of rheumatic cardiac disease is not more than 13 years from onset to death it seems a fact, nevertheless, that 10% lived more than 30 years, 25% more than 17 years, and 50% as long as 9 years. There was no essential difference between the sexes.

The point has already been made that about two-thirds of all cases of rheumatic cardiac disease began in childhood. It is important to know that in spite of an early beginning 5% survived into the later periods of life (46 years and beyond). One-third which began at this period did not live longer than childhood (12 years in females and 14 in males); another third did not survive beyond 18 and 20 years and about 10% did not survive early and middle maturity. In adolescence, which is taken to end at 18 and 20, about 10% of cases began, but only 15 and 19% died within this period. The larger number went on to realize a fairly long expectation of life, even to advanced age.

Recurrences take place at all ages, but are to be encountered most frequently before the age of 20, the maximum being from about 5 to 14. From 14 they taper off to 20 but even after that there is a reasonable number. Before 13 there are more cases among girls than in boys — afterwards there seems to be little difference.

A matter of interest is what may be called the persistence of type, the types being designated carditis, sub-acute activity, polyarthritis, muscle and joint pains, chorea and a type without manifest infection. Each can occur and persist as a pure type. What this study has brought to light, carried on in individual cases over so long a period of time, is the possibility of dividing sufferers into those which exhibit pure types as against mixed forms. When the onset is before 30, polyarthritis is the most common pure form. It increases in frequency from childhood to early maturity. Carditis is much less frequent as a pure form in childhood but becomes somewhat more frequent in the next two periods (adolescence and early maturity). The mixed forms, consisting of two or more types, are commonest in childhood, but decrease later on. When the forms are mixed the commonest arrangement is for carditis and polyarthritis to co-exist. But conversely pure forms increase with age. A matter not without interest is the fact that in childhood relatively speaking few cases can be grouped as being without manifest infection. But after 30, they may amount to as many as a half. These remarks suggest the generalization that aside from persistence of type there is an age distribution of types; that at all ages polyarthritis as a pure type is most common, but that it becomes somewhat less important later and that a part of its place is taken by carditis. To be without manifestation is rare in childhood but increases up to 50% in advanced age.

Mention has already been made of the fact that if the disease begins in a mild fashion its course is likely to be mild. If in childhood the onset is severe, most cases have come to an end before the end of adolescence (80%). Similar general remarks can be made when the onset is later, in adolescence or in maturity. When the onset in childhood is mild almost 40% survive to the end of the period of early maturity and at least 10% attain late maturity. Similar statements can be made concerning mild cases when these are acquired at later periods.

Experience has been gained of certain clinical manifestations. One of them is fibrillation of the auricles. The longer the disease lasts the more likely is auricular fibrillation to occur. If it lasts less than 4 years only 20% exhibit this cardiac irregularity. If it lasts longer than 30 years, 72% do. Clearly, since other occasions for bringing about auricular fibrillation tend to exist as life goes on it is not certain that rheumatic infection is the sole occasion for this appearance. But the increase in numbers affected is significantly uniform with lengthening in the duration of the disease. It would be a matter of very great interest to know what underlies this phenomenon. For the moment no explanation can be offered: whether the size of the heart, intoxication or anatomical lesions are at the bottom of this mischief.

It is striking that although most cases are found in young persons this is the period at which, although cases exist, there are fewest of auricular fibrillation. What the prognostic chances of this occurrence are can be known with reasonable certainty. If it occurred in childhood not more than 10% survived three years; if it began in adolescence, about 10% survived four years; when it began after 20, 10% survived 7 years. Many people at these later ages went on 14 years and more. Put in another way, about 30% of those over 20 survived beyond 3 years. Stated in still another way, children (80%) died in less than a year after the onset of auricular fibrillation; adolescents (80%) survived just a little more than 2 years; and in maturity, death did not take place (80%) until after 4½ years. In short, death occurs sooner after the beginning of auricular fibrillation in childhood and is much longer delayed in older people.

In general clinical manifestations change with age. The number of severe cases is greatest in childhood and declines conspicuously thereafter. But cases of diminished functional capacity are fewer before the age of 30 and this is the case also in congestive heart failure. After this period there are more. Multiple valvular affections, aortic and mitral, are common enough at all ages but are greatest in number in adolescence. The mitral valve is much more frequently involved than the aortic — the number of the aortic cases occurring alone (aortic insufficiency) is infrequent, being greatest in adolescence. Auricular fibrillation, as has been pointed out, relatively infrequent (5%) in childhood, becomes frequent (75%) after 46 years.

The question is often raised — how do patients suffering from rheumatic cardiac disease die? Briefly, about 90% succumb to some form of death traceable directly to an affection of the heart. Rheumatic activity including cases in which congestive failure is also present accounts for about 30%; subacute bacterial endocarditis for less than 10%; congestive heart failure for about 30%; unspecified heart disease, about 20%. A scattering number died of embolism, cerebral accident and unknown cardiac causes. Less than 5% died of causes not related to the heart or circulation.

From these studies the following conclusions can be drawn concerning the natural history of this disease, in this locality. The sexes are about equally afflicted. Particular forms of valvular involvement are not associated with one or the other sex. There is a relation between the period of fairly good health and the relatively speaking short period of rapid decline and death. Auricular fibrillation occurs infrequently in childhood and more frequently in advancing years. Although there are cases in which purity and persistence of type prevail, mixed forms nevertheless are not the exception but the rule. In childhood mixed forms are the more common but afterwards the pure forms.

These studies could not have been carried out without the assistance of many physicians in the cardiac clinics in New York, the help and advice of the Research Committee of the New York Heart Association and the constant and intelligent devotion of the staff.

Alfred E. Cohn, M.D.
Miss Claire Lingg
New York, N. Y.